CLAIMS

1. A genetically transformed plant able to produce a lysosomal enzyme of animal or human origin, characterised in that:

- said plant is transformed via the use of an expression vector comprising:

a. a promoter of a plant gene specific for the expression in seed storage organs and stage-specific;

b. a DNA sequence encoding the signal sequence of a plant protein able to dispatch said lysosomal enzyme to seed storage organs and to provide the post-translational modifications required for the expression of the enzyme in active form;

c. a DNA sequence encoding said lysosomal enzyme deleted of the native signal sequence;

- said enzyme is expressed in seed storage tissues in enzymatically active form and in an amount of at least the 0.8% of the total proteins of the seed.

2. The plant according to claim 1, characterised in that the expression vector is a plasmid.

3. The plant according to claims 1 or 2, characterised in that the promoter derives from the gene of 7S soy globulin.

4. The plant according to any one of the claims 1 to 3, characterised in that the DNA sequence encoding the signal sequence derives from the gene of the 7S soy globulin and is fused to the sequence encoding the structural portion of the mature lysosomal enzyme deleted of the native signal sequence.

5. The plant according to any one of the claims 1 to 4, wherein the lysosomal enzyme expressed in enzymatically active form in seed storage tissues is:

α-N-acetylgalactosaminidase, acid lipase, aryl sulfatase A, aspartylglycosaminidase, ceramidase, α-fucosidase, α-galactosidase A, β-galactosidase, galactosylceramidase, glucocerebrosidase, α-glucosidase,

β-glucuronidase, heparin N-sulfatase, β-hexosaminidase, iduronate sulfatase, α-L-iduronidase, α-mannosidase, β-mannosidase, sialidase and sphingomyelinase.

6. The plant according to any one of the claims 1 to 5, wherein said plant is a *Leguminosa*, cereal, or tobacco.

7. A method for producing the genetically transformed plant able to produce a lysosomal enzyme according to any one of the claims 1 to 6, characterised in that:

- plant cells are transformed via the use of an expression vector comprising:

a. a promoter of a plant gene specific for the expression in seed storage organs and stage-specific;

b. a DNA sequence encoding the signal sequence of a plant protein able to dispatch said lysosomal enzyme to seed storage organs and to provide the post-translational modifications required for expression of the enzyme in active form;

c. a DNA sequence encoding said lysosomal enzyme deleted of the native signal sequence;

- said cells are used to regenerate said transformed plant.

8. The method according to claim 7, wherein said plant is a *Leguminosa*, cereal, or tobacco.

9. A seed of genetically modified plant able to express a lysosomal enzyme, characterised in that:

- said seed contains an expression vector comprising:

a. a promoter of a plant gene specific for the expression in seed storage organs and stage-specific;

b. a DNA sequence encoding the signal sequence of a plant protein able to dispatch said lysosomal enzyme to seed storage organs and to provide the post-translational modifications required for the

expression of the enzyme in active form;

c. a DNA sequence encoding said lysosomal enzyme deleted of the native signal sequence;

- said enzyme is contained in seed storage tissues in enzymatically active form and in the amount of at least the 0.8% of the seed total proteins.

10. The seed according to claim 9, characterised in that the expression vector is a plasmid.

11. The seed according to claims 9 or 10, characterised in that the promoter derives from the gene of 7S soy globulin.

12. The seed according to any one of the claims 9 to 11, characterised in that the DNA sequence encoding the signal sequence derives from the gene of 7S soy globulin and is fused to the sequence encoding the structural portion of the mature lysosomal enzyme deleted of the native signal sequence.

13. The seed according to any one of the claims 9 to 12, characterised in that the lysosomal enzyme expressed in enzymatically active form in seed storage tissues is:

α-N-acetylgalactosaminidase, acid lipase, aryl sulfatase A, aspartylglycosaminidase, ceramidase, α-fucosidase, α-galactosidase A, β-galactosidase, galactosylceramidase, glucocerebrosidase, α-glucosidase, β-glucuronidase, heparin N-sulfatase, β-hexosaminidase, iduronate sulfatase, α-L-iduronidase, α-mannosidase, β-mannosidase, sialidase and sphingomyelinase.

14. The seed according to any one of the claims 9 to 13, wherein said seed is of a *Leguminosa*, cereal or tobacco.

15. A method for producing the seed according to any one of the claims 9 to 14, characterised in that:

- plant cells are transformed via the use of an expression vector comprising:

a. a promoter of a plant gene specific for the expression in seed storage organs and stage-specific;

b. a DNA sequence encoding the signal sequence of a plant protein able to dispatch said lysosomal enzyme to seed storage organs and to provide the post-translational modifications required for expression of the enzyme in active form;

c. a DNA sequence encoding said lysosomal enzyme deleted of the native signal sequence;

- said cells are used to regenerate transformed plants able to produce said seeds.

16. The method according to claim 15, wherein said seed is of *Leguminosa*, cereal or tobacco.

17. A method for extracting and purifying the lysosomal enzyme in active form contained in the seed according to any one of the claims 9 to 14, characterised in that:

a. said seed is ground in liquid nitrogen in the presence of an extraction buffer;

b. the resulting solution is centrifuged;

c. the supernatant is recovered and filtered with filters having a porosity suitable to the enzyme dimensions;

d. the partially purified enzyme is further purified by HPLC chromatography.

18. A use of the seed according to claims 9 to 14, for the preparation of medicaments for enzyme replacement therapies.

19. The use of the seed according to claim 18 for the preparation of a medicament for an enzyme replacement therapy in Gaucher disease.

20. The use of the seed according to claim 18 for the preparation of a medicament for an enzyme replacement therapy in Anderson-Fabry disease.

21. The use of the seed according to claim 18 for the preparation of a medicament for an enzyme replacement therapy in Pompe disease.

22. The use of the seed according to any one of the claims 9 to 14 as means for storing and preserving a

lysosomal enzyme in enzymatically active form.